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This document provides a framework for core competencies in pediatric heart failure. These competencies are built upon the existing ISHLT Guidelines for the Management of Pediatric Heart Failure published in 2014 (ISHLT Monograph Series Volume 8) which received ISHLT Standards and Guidelines approval. We have sent the document for review by the Scientific Council on Pediatric Thoracic Transplant and Heart Failure Council, as well as the Scientific Councils on Heart Failure and Transplantation, Mechanical Circulatory Support, Infectious Diseases, Nursing, Health Science and Allied Health, Pathology, Basic Science and Translational Research, Junior Faculty and Trainees, and Pharmacy and Pharmacology. We have the full support of the Pediatric Thoracic Transplant and Heart Failure Council, Heart Failure and Transplantation Council, Mechanical Circulatory Support Council, Nursing, Health Science and Allied Health Council, and Pharmacy and Pharmacology Council.

The Pediatric Heart Failure Core Competency Curriculum Document is in line with the strategic direction of the Society – namely enhancing the membership value by embracing heart failure as well as transplantation, engagement of our worldwide community. There is expanding interest in the field of pediatric heart failure demonstrated by the creation of the ISHLT Pediatric Heart Failure Guidelines, the newly established ISHLT International Pediatric Heart Failure Registry, and the exceptional attendance at the heart failure session during the ISHLT Annual Meetings.

The curriculum set forth in this document provides a comprehensive framework and the core knowledge for fellows in pediatric heart failure. It also enables established programs to assess their knowledge gaps and provides a framework on which new programs can build. It will serve to establish ISHLT as the leading community for pediatric heart failure knowledge and training and encourage collaborative research and innovation.

In summary, this document represents a succinct but complete framework of the knowledge needed for a pediatric heart failure specialist that is aligned with ISHLT’s recently published Pediatric Heart Failure Guidelines. This document will help lead to the development of an ISHLT Academy Core Competency Course that will be of significant interest to ISHLT members as well as throughout the entire pediatric cardiology community.

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2. To understand the current classification systems for heart failure severity and disease staging

A. Clinical symptoms of heart failure
   • Poor growth and feeding difficulties
   • Respiratory distress
   • Exercise intolerance, fatigue
   • Nausea/vomiting

B. Clinical signs of heart failure
   • Tachycardia
   • Respiratory abnormalities
   • Diaphoresis
   • Hepatomegaly
   • Vital Sign Abnormalities
   • Jugular venous distention
   • Peripheral Edema
   • Delayed capillary refill
   • Metabolic acidosis
   • Altered mental status
   • Other signs of heart failure

C. Heart Failure Severity
   • New York Heart Association Classes (NYHA I-IV)
   • Ross classification (old and new)
   • Intermacs profile
   • Children vs adults

D. Disease Staging
   • American Heart Association Class A-D

References 4-10

II. Epidemiology, Natural History and Prognosis of Heart Failure

Learning Objectives:
1. To understand the incidence, presentation and natural history of the different forms of pediatric cardiomyopathy.
2. To understand the diagnosis, pathophysiology and treatment of the different forms of heart failure that occur in the setting of biventricular and univentricular congenital heart disease
3. To understand the diagnosis, pathophysiology and treatment of the different forms of heart failure due to acquired heart disease in children

A. Heart Failure with Cardiomyopathy
   - Dilated Cardiomyopathy
   - Hypertrophic Cardiomyopathy
   - Restrictive Cardiomyopathy
   - Left Ventricular Noncompaction Cardiomyopathy
   - Arrhythmogenic Right Ventricular Cardiomyopathy

B. Heart Failure due to Congenital Heart Disease
   - Heart Failure in Biventricular Congenital Heart Disease
   - Pre-operative heart failure in volume overload
     • Left to right shunt lesions
     • Valvar regurgitation
   - Pre-operative heart failure due to pressure overload
     • Right heart obstructive lesions
     • Left heart obstructive lesions
     • Congenitally Corrected Transposition (L-TGA)
   - Single Ventricle (pre-operative)
   - Single Ventricle (Stage 1)
   - Single Ventricle (Stage 2)
   - Single Ventricle (Stage 3)
   - Systemic Right Ventricle
   - Other post-operative heart failure

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   - Rheumatic Fever
   - Endocarditis
   - Kawasaki Disease

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III. Etiology and Pathophysiology of Heart Failure

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1. To understand myocyte structure and function as well as basic ventricular mechanics and interventricular interactions
2. To understand the pathophysiology and clinical presentation of the different types of heart failure

A. Myocyte Structure and Contractility
   - Composition of cardiac muscle fibers and sarcomeres
   - Myocardial development
   - Junctions and desmosomes
   - The role of calcium in myocardial contractility

B. Systolic Heart Failure (Heart Failure with reduced EF)
   - Frank Starling Mechanism
   - Pressure-Volume Loops
C. Diastolic Heart Failure (Heart Failure with preserved EF)
   • Preserved systemic ejection fraction
   • Reduced systemic ejection fraction
   • Pressure Volume Loops

D. Biventricular Interactions in Heart Failure
   • Dyssynchrony
   • Ventricular Cross-Talk

E. Clinical Manifestations of Heart Failure
   • Signs/symptoms in infants
   • Signs/symptoms in children and adolescents

F. Genetics of Cardiomyopathy
   • Genetic Testing
     • Hypertrophic Cardiomyopathy
     • Dilated Cardiomyopathy
     • Arrhythmogenic Right Ventricular Cardiomyopathy
     • Restrictive Cardiomyopathy
     • Left Ventricular Noncompaction Cardiomyopathy
   • Variants of unknown significance

G. Heart failure in Congenital Heart Disease
   • Left to right shunt lesions
   • Pressure overload
   • Volume overload
   • Myocardial ischemia
   • Single ventricle failure
     • Ventricular dysfunction
     • Atrioventricular valve regurgitation
     • Systemic-pulmonary collateral vessels
     • Sinus node dysfunction and arrhythmias
     • Pulmonary venous obstruction
     • Protein losing enteropathy
     • Plastic bronchitis

References 158-384

IV. Diagnostic Approach to Pediatric Heart Failure

Learning Objectives
1. To understand available modalities for the diagnostic evaluation of children with newly discovered heart failure.
2. To understand the monitoring of chronic heart failure and the role of various diagnostic tests in care of children with significant systolic and/or diastolic cardiac dysfunction
3. To review the advantages, disadvantages, scope, limitations, risks and contraindications of the various modes of cardiac testing commonly used in children.
4. Review current guidelines on the timing and frequency of cardiac diagnostic testing for children with acute or chronic heart failure.
A. Diagnostic Tests
   • Modalities of cardiac testing
   • General diagnostic testing
   • Basic laboratory profiles
   • General approach to testing
     • New diagnosis
     • Longitudinal management
   • Special considerations in special populations

B. B-type Natriuretic Peptide
   • Chemistry, source, and production of BNP
   • Role in adult heart failure
   • Role in pediatric heart
   • Other biomarkers

C. Cardiac Imaging
   • Chest radiography
   • Electrocardiogram
   • Echocardiography
     • Transthoracic
     • Transesophageal
   • Cardiac MRI
     • Role in cardiomyopathies
     • Role in Myocarditis
     • Measures of ventricular function
     • Role of Stress MRI
     • Risks and deficiencies of MRI

D. Role of Exercise Testing
   • Evaluation of exercise capacity

E. Morbidity & Mortality with certain diagnostic testing

F. Sleep study
   • Central and obstructive sleep apnea assessment in heart failure
   • Hypopnea in heart failure

G. Ambulatory Electrocardiographic Monitoring
   • Assessment for arrhythmia
   • Monitoring medications effects

H. Cardiac Catheterization and Endomyocardial Biopsy
   • Assessment of hemodynamics
   • Hemodynamics and relation to outcomes
   • Role of angiography in assessment
   • Role of endomyocardial biopsy
   • Assessment of pulmonary vascular resistance (PVR)
     • Calculation of PVR
     • Assessment of reactivity
     • Monitoring of PVR in the listed patient

I. Invasive Electrophysiologic Testing
   • Evaluation for arrhythmia
   • Risk stratification
     • Preparation for placement of pacemakers and implantable
Learning Objectives:
1. To understand models that explain the pathogenesis for the progression of heart failure.
2. To be able to identify how the various steps in the pathogenesis of heart failure are being targeted by medications currently available, medications that have failed, and medications that are being designed.
3. To be able to synthesize how various combinations of medical therapies can improve outcomes.
4. To be knowledgeable on what is recommended with a high level of evidence by adult guidelines and whether and how they may apply to children given available Pediatric and Congenital Heart Disease Guidelines and available published literature.
5. To understand significant adverse effects and possible contraindications of different heart failure medicines
6. To understand within each class of heart failure medications the possible clinical scenarios that would lead to different choices of medications
7. To be knowledgeable on drug to drug interactions and how they may influence medication management.
8. Recognize the differences in metabolism and pharmacokinetics in pediatrics

A. Symptomatic Relief of Heart Failure
   • Diuretics
   • Vasopressin receptor antagonists
   • Nesiritide

B. The renin-angiotensin-aldosterone axis
   • Angiotensin converting enzyme inhibitors
   • Angiotensin receptor blockers (ARB)
   • ARB plus neprilysin inhibitor
   • Mineralocorticoid receptor antagonists

C. The sympathetic nervous system
   • Beta blockers
   • Digitalis
   • Ivabradine

D. Other
   • Isosorbide dinitrate plus hydralazine
   • Phosphodiesterase 5 inhibitors

E. Inotropes
   • Short-term use
   • Long-term use
   • Dobutamine, Dopamine, Epinephrine, Milrinone, Levosimendan
   • Other Inotropes

F. Drug to drug interactions and drug monitoring
G. Primary and secondary prevention of arrhythmias
H. Primary and secondary prevention of thromboembolic complications
   • Anti-platelets
   • Anti-coagulants
   • Clinical trials of thromboembolic prevention in heart failure
I. Novel medical therapies in the horizon
   • Stem cell therapy
   • Others
   • Pharmacogenomics
J. Prevention of dilated cardiomyopathy and heart failure
   • Treatment in asymptomatic heart failure (NYHA I) or Stage A heart failure (pre-clinical disease)

References 755-925

VI. Pharmacological Treatment Of Heart Failure With ‘Preserved' EF

Learning Objectives:
1. To clearly define the terminology that has evolved from diastolic heart failure to heart failure with preserved ejection fraction in the adult population, in order to properly apply the published knowledge to pediatric and congenital patients.
2. To understand the pathogenesis of HFpEF and diastolic heart failure
3. To understand cardiac diastolic physiology

A. Therapies with Class I Indications
   • Diuretics
   • Renal monitoring
   • Blood pressure control
   • Dietary modifications
B. Therapies with Class II Indications
   • ACE inhibitors and angiotensin receptor blockers
   • Calcium channel blockers
   • Mineralocorticoid/aldosterone receptor antagonists
   • Phosphodiesterase inhibitors
C. Therapies with Class III Indications
   • Digoxin and other digitalis glycosides
   • Positive inotropic agents
   • Pulmonary vasodilators

References 926-956

VII. Electrophysiology in Heart Failure

Learning Objectives
1. To recognize the indications for pacemaker therapy in pediatric heart failure
2. To recognize the indications for cardiac resynchronization therapy in pediatric heart failure
3. To recognize the indications for ICD therapy in pediatric heart failure
4. To recognize the indications for electrophysiology study and ablation therapy in the presence of cardiomyopathy

A. Pacemaker therapy
   • Indications in pediatric heart failure
   • Management
   • Complications in heart failure

B. Cardiac Resynchronization
   • Indications in pediatric heart failure
   • Diagnostic criteria for dyssynchrony

C. Cardioversion & defibrillator therapy
   • Indications in pediatric heart failure
   • Automated external defibrillators (AED’s)
   • Implantable cardiac defibrillators (ICD’s)
   • Adverse events

D. Electrophysiologic Testing and Ablation Therapy
   • EP testing in setting of ventricular dysfunction
   • Ablation for tachycardia-induced cardiomyopathy
   • Ablation for ventricular arrhythmia and cardiomyopathy

References: 957-1026

VIII. Surgical Treatment of Pediatric Heart Failure

Learning Objectives:
1. To understand the history of mechanical circulatory support and be familiar with older devices and why they were not successful
2. To understand the indications for the use of MCS in children and which devices are available and how these devices can be used
3. To understand special circumstances in pediatric MCS such as biventricular support, support in the single ventricle population and the role for destination therapy in children.
4. Review devices in development and future avenues for MCS in pediatrics

A. Overview of Mechanical Circulatory Support

B. History and development of ventricular support systems
   • Durable Mechanical Circulatory Support
     • Goals of support
     • Indications
     • Patient Selection
     • Timing of implantation
     • Current devices
       • Berlin Heart Excor
       • HeartMate II
       • Heartware HVAD
       • Syncardia Total Artificial Heart
   • Temporary Mechanical Circulatory Support
     • Rotaflow
• Centrimag/Pedimag
• Impella
• Balloon Pump
• Extracorporeal Membrane Oxygenation

C. Special Topics
• Biventricular support (BiVAD’s)
• Right ventricular failure after LVAD
• MCS for restrictive and hypertrophic cardiomyopathy
• MCS in the univentricular heart
• Recognizing recovery and timing of device explant
• Destination Therapy in pediatrics
• Anticoagulation
• Adverse events

D. Future Directions

References: 1027-1104

IX. Co-morbidities

Learning Objectives
1. To understand the pathophysiology of the co-morbidities frequently found with pediatric heart failure
2. To understand the best treatment options for these medical co-morbidities to maximize outcomes
3. To understand the importance of a psychological evaluation and physical activity in pediatric patients with heart failure.

A. Anemia
• Risk factors, etiologies, diagnosis
• Erythropoiesis-stimulating agents
• Use of iron suppletion

B. Renal Dysfunction
• Markers of renal function
• Evaluation of creatinine clearance and glomerular function
• Pediatric RIFLE scoring system

C. Airway and Parenchymal Respiratory Morbidity
• Interaction of cardiac dysfunction and lung mechanics
• Sleep disordered breathing
• Airway abnormalities (congenital and acquired)
• Use of bronchodilators and inhaled corticosteroids
• Pulmonary function testing

D. Infectious Diseases In Heart Failure
• Risk of respiratory and gastrointestinal illnesses
• Use of palivizumab for prevention of respiratory syncytial virus (RSV)
• Anti-viral therapy for treatment of respiratory infections
• Adjustment of diuretics in gastrointestinal illness
• Immunization administration/timing
• Infections related to Mechanical Circulatory Support
E. Malnutrition and Cachexia
   • Enteral nutrition supplementation
   • Nasogastric and gastric feeding tubes
   • Intravenous nutrition
   • Electrolyte monitoring and supplementation
   • Vitamin and mineral supplements

F. Metabolic Syndrome
   • Hypertension
   • Dyslipidemia
   • Insulin resistance
   • Kidney disease
   • Liver disease

G. Depression & Psychological Functioning in Pediatric Heart Failure
   • Mood disorders (depression and/or anxiety)
   • Effects of ICD placement and inappropriate shocks
   • Adjustment disorders
   • Sleep disorders

H. Cognitive & Psychosocial Performance in Pediatric Heart Failure
   • Impaired cognitive development (intelligence, motor, speech/language)
   • Impaired social development
   • Congenital heart disease vs non-congenital heart disease
   • School performance and failure
   • Early intervention programs and developmental therapies
   • Referral to developmental specialists

I. Exercise Training and Activity Recommendations
   • Pre-participation screening
   • Risk stratification
   • Exercise testing (>6-8 years old)

References 1105-1272

X. Acute Heart Failure

Learning Objectives:
1. To recognize the clinical presentation of acute heart failure
2. To understand the pathophysiologic process that leads to acute heart failure
3. To learn appropriate management and treatment of acute heart failure

A. Definition, Etiology and Epidemiology
   • Definition of acute heart failure
   • Etiology of acute heart failure
   • Epidemiology
   • Initial assessment
      • Heart failure due to ventricular dysfunction
      • Heart failure due to volume overload
      • Heart failure due to pressure overload
B. Patient Monitoring
   • Hemodynamic monitoring
     • Invasive
     • Non-invasive
   • Laboratory monitoring
C. Evaluation After Stabilization
   • Medical therapy with chronic inotropes
   • Continuous positive pressure ventilation
     • Non-invasive positive pressure ventilation
     • Invasive positive pressure ventilation
   • Indication and timing for mechanical support
   • Evaluation for heart transplantation
D. Treatment of Acute Heart Failure
   • Goal directed therapies
   • Optimizing oxygen delivery
     • Preload
     • Afterload
     • Contractility
   • Decreasing oxygen consumption
E. Fluid Management
   • Optimize preload
   • Avoid Overload
F. Nutrition
   • Optimize caloric intake
G. Considerations in the Treatment of Acute Right Heart Failure
   • Management of right heart dysfunction
   • Role of pulmonary vasodilators
H. Perioperative Acute Heart Failure
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   • Medical vs surgical Treatment

References: 1273-1464

XI. Special Patient Populations

Learning Objectives
1. To understand the similarities and differences in clinical presentation of heart failure in special patient populations
2. To learn specific management and interventional strategies available for heart failure treatment in special patient populations
3. To understand the pathophysiological processes that lead to heart failure in special patient populations

A. Secondary Right Ventricular Failure
   • Pulmonary Hypertension
   • Pulmonary Embolism
   • Right Ventricular Volume Overload
B. Systemic Right Ventricular Failure in a Biventricular Circulation
   • Traditional Heart Failure Medical Management
   • Medical Management specific to the right ventricle
   • Cardiac resynchronization therapy (CRT)
     • Evaluation for CRT candidacy
     • Lead placement in CRT
   • Surgical Options
   • Mechanical Circulatory Support

C. Single Ventricle Heart Failure
   • Perioperative Heart Failure
   • Early Fontan Failure
   • Chronic single ventricle heart failure
     • Possible medical interventions
     • Possible surgical interventions
     • Possible catheter interventions

D. Neuromuscular Disorders
   • Clinical Cardiac Evaluation
   • Dystrophinopathies
   • Limb-Girdle muscular dystrophies
   • Laminopathies
   • Myotonic dystrophies
   • Freidreich ataxia
   • Barth Syndrome
   • Storage Disorders

E. Cancer and Heart Failure
   • Mechanism of cardiotoxicity
   • Clinical presentation
   • Chemotherapy Induced Cardiomyopathy
     • Screening and Prevention
     • Treatment and Prognosis

F. Adult Heart Failure & Congenital Heart Disease
   • Medical therapies
   • Surgical therapies for adult congenital heart disease (ACHD)
   • Mechanical circulatory support for adults
   • Transplantation in ACHD
   • Ex vivo organ perfusion

References: 1465-1578

XII. Transplantation

Learning Objectives:
1. To understand the indications for pediatric heart transplantation for pediatric heart failure
2. To recognize appropriate timing for referral to a heart transplant team
3. To understand the evaluation process
4. To understand risk factors for poor outcome following heart transplantation
To understand unique risk factors in end stage congenital heart disease

To understand how tools that predict transplant outcomes can aide in candidate selection and in counseling families

To describe current outcomes for pediatric heart transplantation

To understand causes of heart failure in the transplanted heart

A. Indications and Referral
   • Brief history of pediatric heart transplantation
   • Indications
     • Age-specific indications
     • Myocarditis
     • Dilated cardiomyopathy
     • Congenital heart disease, unoperated
     • Congenital heart disease, rescue
     • Impact of systemic disease
     • Restrictive Cardiomyopathy
     • Hypertrophic Cardiomyopathy

B. Evaluation Process
   • Multi-disciplinary team

C. Specific Risk Factors
   • Pulmonary hypertension
   • HLA sensitization
   • End organ function
     • Renal
     • Pulmonary
     • Hepatic
     • Neurologic
   • Infection
   • Malignancy
   • Cachexia/Obesity
   • Psychosocial
   • Special considerations in congenital heart disease
   • Prior sternotomy
   • Pulmonary artery and pulmonary vein anatomy
   • Pulmonary AVM and other collaterals

D. Pre-transplant Risk Prediction
   • Use of risk models to predict survival

E. Indications for Heart Transplantation
   • Current guidelines
   • Ethical dilemmas and contraindications to transplant

F. Overall Outcomes
   • Survival
   • Complications
     • Primary graft failure
     • Acute rejection
     • Coronary allograft vasculopathy
     • Post-transplant lymphoproliferative disease
     • Renal dysfunction
• Quality of life

G. Heart Failure Post Transplant
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H. Re-Transplantation
   • Indications

References: 1579-1672

XIII. Health Care Delivery

Learning Objectives:
1. To learn the importance of a standardized management system for delivering effective heart failure care
2. To learn the components of discharge planning necessary to prevent readmissions after an acute heart failure inpatient stay
3. To learn the components of an effective outpatient pediatric heart failure management program
4. Recognize the importance of a multidisciplinary team approach to provide comprehensive pediatric heart failure care

A. Overview
B. Disease Management Systems
   • Readiness for discharge
   • Transition to home
   • Working with local practitioners
   • Multidisciplinary Management Programs
      • The Heart Failure Clinic
      • Home Based Heart Failure Management Program
      • The Self-Physician Oriented Program
   • Remote monitoring for home surveillance

C. Transition from Pediatric to Adult Care
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      • Timing of Transition

D. Palliative Care & End of Life Support
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      • Role in pediatric heart failure
      • Team members
      • Introduction of palliative care team
      • Collaboration with heart failure team
      • Quality of Life Focus

E. Standardization of Practice Guidelines
   • Metrics for outpatient pediatric heart failure care
XIV. Research in Pediatric Heart Failure

Learning Objectives
1. Recognize the different types of research
2. Understand the most common statistical methods used in clinical research
3. Recognize the challenges to performing research in pediatrics

A. Types of research
   • Clinical research

B. Types of research studies
   • Basic science research
   • Translational research

C. Basic biostatistics
   • Types of data
   • Measures of central tendencies
   • Types of distribution
   • Sample size and Power of study

D. Critical appraisal of published research

E. Challenges in pediatric research

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